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Rapid progression of tracheal stenosis associated with tracheopathia osteo-chondroplastica

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Abstract. Tracheopathia osteochondroplastica (TPO) is a rare, but increasingly recognised condition in which there is accumulation of calcium phosphate with benign submucous proliferation of cartilage and bone beneath the tracheal mucosa, often with squamous metaplasia of the mucosal columnar epithelium. This condition is usually asymptomatic, but may be slowly progressive, causing haemoptysis, dry cough and dyspnoea. We report a case of TPO in which there was rapid progression of tracheal stenosis such that the size of endotracheal tube that the upper airway would accept changed from 8.00 mm to 3.0 mm during a six-week period. This extreme reduction in airway calibre had not been detected on spirometry nine days prior to his final admission. This is the first report of such rapid progression of tracheal stenosis associated with TPO.

Key words: Tracheopathia osteochondroplastica – Tracheal stenosis

Case history

A 54-year-old man who weighed 106 kg was admitted from the Casualty Department in acute respiratory failure. He had a one-week history of a cough productive of 'grey' sputum and worsening shortness of breath. He had an unremarkable past medical history with the exception of episodes of acute bronchitis for three years following 'flu like' illnesses. Two weeks prior to admission, his general practitioner commenced bronchodilator therapy. He had smoked 15 cigarettes a day for thirty years. Social and family history were unremarkable.

On examination, he was obese, drowsy and unable to obey commands. He was centrally cyanosed with cold clammy peripheries, a regular pulse rate of 100/min, a blood pressure 80/60, and a jugular venous pressure elevated to the angle of the jaw. Oedema of his ankles was present. Heart sounds were normal with no added sounds. There were no abnormal findings elsewhere. Arterial blood gas (ABG) analysis revealed PO_2 7.2 kPa, PCO_2 12.1 kPa and pH 7.23. He was intubated on arrival on the Intensive Care Unit (ICU) with an 8.00 mm endotracheal tube (Portex) and ventilated on the Servo 900B ventilator.

Peak inflation pressures were 30 cm H_2O. Intubation was difficult due to a short neck and thick set jaw – but a good view of the vocal cords was obtained.

Initial investigations showed, Hb = 14.1 gdl^{-1} (normal indices), WCC = $11.8 \times 10^9 l^{-1}$ (predominant granulocytosis) sodium = 122 $mmol^{-1}$, potassium = 4.7 $mmol^{-1}$, creatinine = 83 $\mu mols^{-1}$, bicarbonate = 29 $mmol^{-1}$. Liver function tests were normal and cardiac enzymes were not elevated.

Chest x-ray revealed normal cardio-thoracic ratio with clear lung fields. The electrocardiogram had a normal axis without any acute changes. There was a heavy growth of haemophilus influenzae in the sputum. Blood cultures were negative.

He made steady progress on the ICU following full ventilation, regular physiotherapy and antibiotic therapy with Cefuroxime 750 mg i.v. tds. He required sedation with Midazolam, but pulled out his endotracheal tube on two occasions with the cuff inflated. He was re-intubated on both occasions with an 8.0 mm tube, which passed with ease into the trachea.

An unsuccessful attempt to wean onto continuous positive airway pressure (CPAP) of 5 cm H_2O was made on day 3 but by day 5 he managed to maintain acceptable ABG of PO_2 = 7.45 kPa, PCO_2 6.5 kPa, pH = 7.37 on CPAP of 5 cm H_2O with a respiratory rate of 20 and F_IO_2 of 0.35. His forced vital capacity (FVC) was 1.2 l. Following extubation he continued to make a steady recovery and he was discharged home

on day 12. Arterial blood gases on discharge were $PO_2 = 7.44$ kPa, $PCO_2 = 4.7$ kPa, pH = 7.39.

On review one month after discharge there was marked improvement in his lung function tests compared to admission values. His FVC had improved from 2.31 at discharge to 3.11 and his forced expiratory volume in one second (FEV_1) from 0.91 to 1.41. The ratio FEV_1/FVC was unchanged at 39, indicating moderate airflow obstruction. His resting oxygen saturation was 90% and $PCO_2 = 5.3$ kPa.

Nine days after review in the clinic he was re-admitted to the ICU following a sudden deterioration into acute respiratory failure, clinically thought to be secondary to a further episode of acute bronchitis. On this occasion, it proved impossible to pass an 8.0 mm ET tube due to an obstruction in the upper airway. Various smaller endotracheal tubes were tried and a 3.0 mm tube could just be passed into the trachea a short distance and held with Magill's forceps, allowing for manual inflation of oxygen.

A decision was made to proceed to an emergency tracheostomy. This proved to be technically difficult due to his obesity and short neck although ventilation appeared adequate. Tracheostomy was eventually performed but he developed a bradycardia followed by ventricular fibrillation and resistant asystole despite all attempts at resuscitation.

Post mortem confirmed that the trachea was greatly narrowed just below the cricoid cartilage (Fig. 1) due to the presence of TPO (Fig. 2). This involved the first three tracheal cartilages. The tracheal mucosa in the stenotic region showed evidence of trauma with replacement by maturing granulation tissue and fibrous tissue (Fig. 2). Evidence of chronic obstructive airways disease and a small primary adenocarcinoma of the lung without secondaries were also noted.

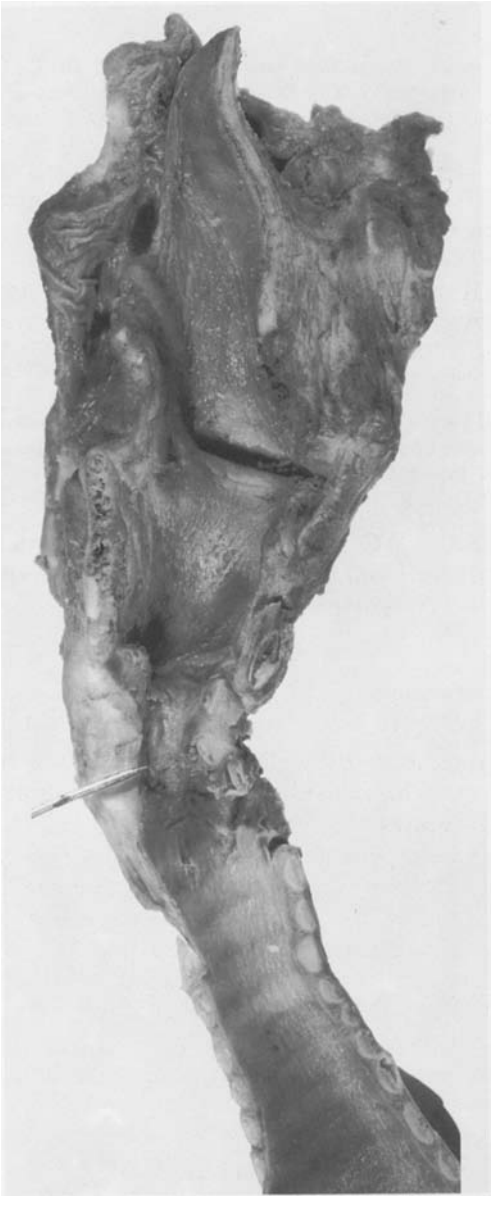

Fig. 1. Larynx and trachea showing hard, apparently confluent irregular nodules, 1.5 cm wide, lying immediately beneath the laryngeal cricoid cartilage, within the cartilaginous part of the tracheal circumference

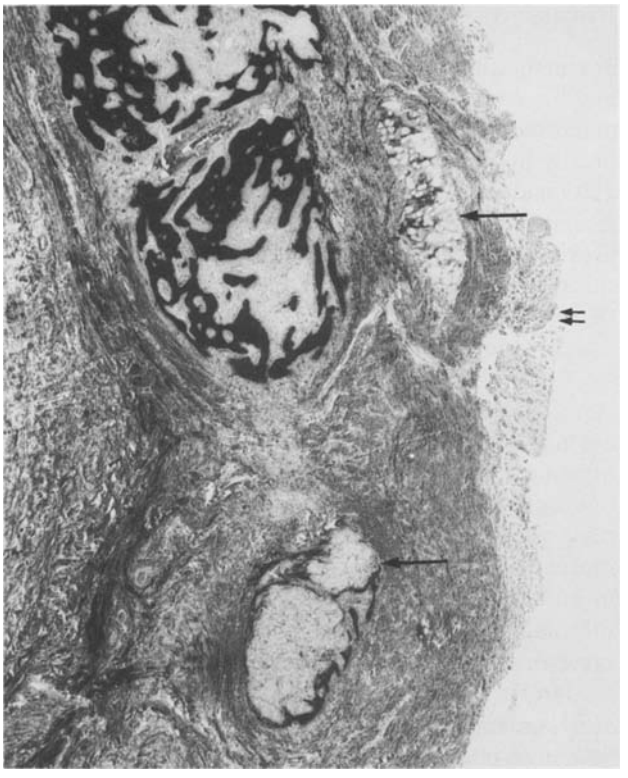

Fig. 2. Two cartilaginous nodules (*single arrows*), surrounded by connective tissue, with the first tracheal cartilage. Granulation tissue maturing into fibrous tissue is also present (*double arrows*). Elastic van Gieson X 14.53

Discussion

Tracheopathia osteochondroplastica (TPO) is a rare but increasingly recognised condition first described macroscopically by Rokitansky in 1855 and microscopically by Wilks in 1857. Until 1974, only 245 cases of TPO had been reported in the literature [1]. The cause remains obscure, although many theories have been proposed. It occurs most commonly in the lower two thirds of the trachea and often extends to involve the first portion of the major bronchi. The larynx is usually spared. The bronchoscopic description of a 'rock garden' in TPO is due to the submucosal accumulation of calcium phosphate and benign enchondroma-like growths of cartilage and bone. The overlying mucosa is usually intact and histologically normal, but squamous metaplasia may be present. There is a male preponderance of 3:1 with the disease usually presenting over the age of 50, but it has been reported in children. Patients are usually asymptomatic with the diagnosis being made at bronchoscopy, but may present with haemoptysis, dry cough or dyspnoea.

TPO is considered to be progressive, but the rate of development of the lesion is unknown. No studies have been published of serial bronchoscopy and tomography in these patients. Clee et al. 1983 [2] described a case of TPO with repeat bronchoscopy at one year after diagnosis in which there was no obvious progression of the pathology. However, Alroy et al. reported a single case demonstrating significant deterioration during an eight-month period of observation [3]. Our case illustrated very rapid progression of tracheal stenosis associated with TPO. The trauma of repeated intubations and extubations on the narrowed and roughened tracheal wall led to granulation and fibrous tissue formation with further narrowing of the tracheal lumen. The lumen had narrowed to such an extent that it would not accept a 3.0 mm endotracheal tube, having freely accepted an 8.00 mm endotracheal tube, six weeks earlier. Spirometry nine days before his second admission had failed to identify the stenosis with the recordings actually showing an improvement in lung function. Spirometry has been shown to be a poor indicator of proximal airway obstruction. Flow-volume loops are preferred for laboratory assessment of major airway obstruction as these become abnormal earlier than the spirometric measurements [4]. However, Berend et al. (1979) in studies on two patients showed that unequivocal evidence of central airway obstruction could not be obtained by the use of flow/volume loops or spirometry [5]. Bronchoscopy or tomography remain the diagnostic procedure of choice.

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